NEUROLOGY

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APPROVED
by the Ministry of Education and Science
of Ukraine as a textbook for students of
higher education establishments — medical
universities, institutes, and academies

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The national textbook presents topical diagnostics of nervous system diseases and the main sections of clinical neurology from the standpoint of classical medicine and current scientific advances. The main topical principles of the structure of the central and peripheral nervous system, symptoms of cranial nerve disorders, issues of neurophysiology and its peculiarities in childhood are discussed in detail. The book outlines the fundamentals of modern diagnostics of nervous diseases and its methods (electrophysiological, ultrasound, computed tomography, biochemical), which make it possible to study the structure and functions of the nervous system, its metabolism, and hemodynamics under conditions of physiology and pathology.

The textbook is intended for students of higher education establishments — medical universities, institutes, and academies, and also interns, neurologists, and family doctors.

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CONTENTS

ABBREVIATIONS

A — Amplitude
ABP — Average Blood Pressure
ABCDE — Airways, Breathing, Circulation, Disabilities, Explosure
ABSEP — Acoustical Brain Stem Evoked Potential
ACE — Angiotensin Converting Enzyme (Inhibitors)
ACEP — Anticholinesterase Preparations
ACTH — Adrenocorticotrophic Hormone
ADCC — Acute Disturbances of the Cerebral Circulation
ADEM — Acute Disseminated Encephalomyelitis
AHE — The Acute Hypertensive Encephalopathy
AIDS — Acquired Immune Deficiency Syndrome
ALS — Amyotrophic-Lateral Sclerosis
ANS — Autonomic Nervous System
APDT — Adsorbed Pertussis-Diphtheritic-Tetanic (vaccine)
APMU — Action Potential of the Motor Units
APTT — Activated Partial Thromboplastic Time
APV — Artificial Pulmonary Ventilation
ARA-II — Antagonists of the Receptors to Angiotensin II
ARC — AIDS-related complex
ARVI (ARVD) — Acute Respiratory Viral Infections (Diseases)
ARS — Acute Radiation Synrome
ART — Antiretroviral Therapy
APTT — Activated Partial Thromboplastin Time
ASA — American Stroke Association
ASEM — Acute Scattered Encephalomyelitis
ACE — Angiotensin-converting Enzyme
ATF — Adenotriphosphoric Acid
AVPU — Alert, Voice, Pain, Unresponsiveness
BAL — British Antilewisite (antidotum)
BBM — Bulbar Biomicroscopy
BI — Brain Injury
BP — Blood Pressure
BPM — Base Protein of Myelin
CC — Concussion, Commotio Cerebri
CA — Coefficient of Absorption
CAG — Cerebral Angiography
CBR — The Complement Binding Reaction
CCA — Common Carotid Artery
CCT — Craniocerebral Trauma
CFT — Complement Fixation Test
CHC — Cerebral Hypertensive Crisis
CHD — Coronary Heart Disease
CJD — The Creutzfeldt—Jakob disease
CNS — Central Nervous System
CP — Cerebral Pasly
CPP — Cerebral Perfusion Pressure
CRD — Chronic Radial Disease
Chr. — Chromosome
CSH — Chronic Subdural Hematoma
CT — Computed Tomography
DBS — Deep Brain Stimulation
DE — Discirculatory Encephalopathy
DICS — Disseminated Intravascular Coagulation Syndrome
DM — Diabetes Mellitus
DNase — Deoxyribonuclease
EAP — Electro Active Potential
Echoes — Echoencephalography
ECG — Electrocardiography
EEG — Electroencephalography
ELISA — Enzyme-Linked Immunosorbent Assay
EMG — Electromyography
EMP — Evoked Motor Potentials
EP — Evoked Potentials
ESR — Erythrocyte Sedimentation Rate
FAG — Fluorescent Angiography
FI — Fatal Insomnia
FNT — Factor of Necrosis of Tumors
FLAIR — Fluid Attenuated Inversion Recovery
FM — Frontal Mastoidal (lead)
FMRT — Functional Magnetic Resonance Tomography
GABA — Gamma-Aminobutyric Acid
HAIT — Hemagglutination Inhibition Tests
HAART — High-Active Anti-Retroviral Therapy
HC — Hypertensive Crisis
HEB — Hematoencephalic Barrier

HIV — Human Immunodeficiency Virus
HLA — Human Leukocyte Antigen
HR — Heart Rate
HT — Hemorrhagic Transformation
ICA — Internal Carotid Artery
ICH — Intracranial Hematoma
ICPH — Intracranial Parenchimatous Hemorrhage
ICrP — Intracranial Pressure
IL — Interleukins
IM — Intramuscularly
INR — International Normalized Ratio
ISCDB — Initial Signs of the Circulatory Deficiency of the Brain
IU — International Units
IV — Intravenously
IVH — Internal Ventricular Hemorrhage
KIU — Kininogenous Inhibiting Units
LBFV — Linear Blood Flow Velocity
LD — Lyme disease
LP — Latent Period
LVBF — The Linear Velocity of the Blood Flow
MBD — The Minimal Brain Dysfunction
MBP — The Myelin Basic Protein
MBT — Micobacterium of Tuberculosis
MHC — Microhemocirculation
MMSE — Mini Mental State Examination
MOG — Myelinoligodendrocytic Glycoprotein
MP-methods — Magnetic Perfusion methods
MRA — Magnetic Resonance Angiography
MRI — Magnetic Resonance Image
MRS — Magnetic Resonance Spectroscopy
MSCT — Multi-slice Computed Tomography
MU — Motor Units
NIHSS — National Institute of the Health Stroke Scale
NMDA — N-Methyl-D-aspartate
NSMN — Neural-Senso-Motor Neuropathies
NMR — Nuclear Magnetic Resonance
NSG — Neurosonography
OCT — Optical Coherence Tomography
OM — Occipital Mastoidal (lead)
OPCs — Organophosphorus Compounds
PAMU — Potential of Activity of the Motor Units
PCR — Polymerase Chain Reaction
PD — Prion Diseases
PEG — Pneumoencephalography
PET — Positron Emission Tomography
PF — Potential of Fibrillations
PLP — Proteolipid Protein
PR — The Precipitation Reaction
PRNP — Prion Protein Gene
PrP^{c} — Normal Isoform of Prion Protein
PrP^{Sc} — Pathlogic Isoform of Prion Protein
PPh — Polyphase
PT — Physiotherapy
PVL — Periventricular Leukomalacia
RIHA — Reaction of Inhibition of Hemagglutination
RPHA — Reaction of Passive Hemagglutination
(R)CFT — Reiter's Complement Fixation Test
REG — Rheoencephalography
RIF — Reaction Immunofluorescence
RIT — Reaction of Immobilization of Treponema
RP — Reaction of Precipitation
RVG — Rheovasography
SAH — Subarachnoidal Hemorrhage
SCT — Spinal Computed Tomography
SGSS — Syndrome of Gersmann-Straussler-Sheinker
SLE — Systemic Lupus Erythematous
SPECT — Single Photon Emission Computed Tomography
ST — Spinal Trauma
TB — Tuberculosis
T-cells — Lymphocytes produced in the Thymus
TCDG — Transcranial Dopplerography
TDCC — Transient Disturbances of the Cerebral Circulation
TDS — Titanic-Diphtheritic Serum
TEL — Tetraethyl Lead
T1 — Lymphocytes of Class 1
T2 — Lymphocytes of Class 2
TEH — Traumatic Epidural Hematoma
TIA — Transient Ischemic Attack
TMS — Transcranial Magnetic Stimulation
TNF — Tumor Necrosis Factor
TSH — Traumatic Subdural Hematoma
UA — Units of Action
UHF — Ultra-High Frequencies
USDG — Ultrasound Dopplerography
US — Ultrasound Study
UVR — Ultra-Violet Radiation
VEP — Visual Evoked Potencials
VEP — Visual Electro Potential
VVD — Vegeto-Vascular Dystony
VHV — Variccela/Herpes Zoster Virus
WHO — World Health Organization

PREFACE

Neurology (from Greek *neuron* — nerve, *logos* — science) — is the science about the human nervous system in health and pathology. It studies anatomy, histology, physiology, and biochemistry of the nervous system as well as pathological processes of the human body, which cause disorders of its functions. **Neuropathology** is a part of neurology, which studies diseases of the nervous system.

As an independent clinical science neuropathology was dedicated in 1862. The important role in this process was played by the French neurologist, Professor of the Paris University *Jean Martin Charcot*, who at that time created and headed the world's first clinic in the hospital Salpetrier near Paris and the Chair of nervous diseases at the University for the patients with neurological disorders. The development of neuropathology as a separate field of medicine in the middle of the 19th century was associated with significant achievements in neuroanatomy, neurophysiology, and neurohistology.

In 1884 as an independent academic subject "The nervous and mental diseases" was included into the curriculum of medical faculties of Russian universities. At the same time the united departments of the nervous and mental diseases were established; the first of which was in the Moscow University; it was headed by A.Y. Kozhevnikov. He was also the author of the first Russian textbook on the nervous and mental diseases for students.

The development of the Ukrainian school was connected with the neurology departments of the nervous and mental diseases at the leading University hospitals in Kiev, Kharkiv, and Odessa. The first departments were established in 1884 at the medical faculty of the Kiev and Kharkiv Universities, where the teaching of neurology was carried out by well-known scientists, professors I.A. Sikorskiy and P.I. Kovalevskiy. Further in these departments fruitfully worked such famous scientists as B.N. Mankivskiy, D.I. Panchenko, N.B. Mankivsky, O.P. Vinnytskiy in the Kyiv University and S.N. Davidenkov, A.M. Greenstein, G.D. Leshchenko, E.G. Dubenko in the Kharkiv University. The third Department of the nervous and mental diseases in Ukraine was established at the Novorossiyskiy University in Odessa in 1905 under the leadership of Professor N.G. Popov. In the same year he founded the Department of neurology at the medical faculty of the Lviv University, the leaders of which in different years were Professor D.I. Panchenko, N.V. Mirtowskiy, and D.I. Proniv.

Over the past 25 years in neurology a huge breakthrough in the diagnosis and study of pathogenesis of the nervous diseases was made. Due to achievements of genetics and neurochemistry, improvement of methods of

neuro-imaging neurology has evolved into an exact science. All of this requires a high level of training specialists in different fields — from family physicians to specialists of the narrow specialization.

This textbook was created as the national for the purpose of optimisation the study of neurology for the medical students of higher educational institutions of Ukraine of the level IV of accreditation. The group of authors consists of leading scientists and teachers of all neurology departments of the Medical Universities of our country.

The textbook has two parts. In the first part there are the main issues of propedeutics of the nervous diseases, anatomical and physiological features of the nervous system, the symptoms and syndromes of its defeat at various levels, the methodology of the topical diagnosis. A separate chapter is devoted to additional research methods of the nervous system, which are described as the traditional neurology and the new methods of diagnosis.

The second part presents etiopathogenesis, clinical picture, diagnostics, treatment and prevention of diseases of the nervous system. This part covers almost all kinds of diseases of the nervous system from the widespread cerebrovascular, demyelinated diseases and diseases of the peripheral nervous system to the rare forms of the neurological pathology — prionic diseases, dermatomyosites, lesions of the nervous system in the case of HIV infection, etc. It clearly demonstrates the modern trends in neurology: the creation of joint scientific and practical directions — cardioneurology, somatoneurology, vertebral neurology etc. Principles of the treatment of neurological diseases are based on the system approach and principles of evidence-based medicine.

At the end of each part there are examples of the test tasks and clinical cases for self-control. Figures, tables and schemes are presented to improve the perception and assimilation of the material. The textbook is written to help the students of higher medical institutions, doctors-interns, clinical residents, neurologists, and specialists of family medicine for learning the basic principles of neurology, and for intensifying fundamental knowledge for the personal improvement in the field of neurology.

PART 1

GENERAL NEUROLOGY

CHAPTER 1

BASIC PRINCIPLES OF STRUCTURE AND FUNCTIONING OF THE NERVOUS SYSTEM

GENERAL PRINCIPLES OF STRUCTURE OF THE NERVOUS SYSTEM

The main structural, functional, genetic and anatomic unit of the nervous system is the *nerves cell* or *neuron* consisting of a body and nervous processes of two types: dendrites and axons (see the Figure 1.1 on the coloured insert).

The main function of neuron is reception, processing of information and conducting irritation to other cells. The receptor endings of the sensitive nervous fibers (receptors) perceive the external and internal stimuli and conduct them in the form of impulses by **dendrites** (afferent nervous processes) to the neuron body.

The axon is a long process, which conducts the nervous impulse away from the body cell and has the corresponding effector ending. There is only one axon in the neuron, and its function is to conduct corresponding impulses by synapses from the neuron body to other neurons or working cells (muscular and glandular).

Bodies of neurons of the central nervous system (CNS) form the gray matter of the brain, and on periphery they form the cerebrospinal and autonomic nodes**.** In the CNS **the nervous fibers** or **processes of neurons** are the basis of the white cerebral matter, and they function as the conductors. In the peripheral part of the nervous system they are the part of roots and nerves and conduct nervous impulses from the center to periphery (*efferent fibers*), and vice versa, from the periphery to center (*afferent fibers*). The nervous fiber consists of the axial cylinder (the actual process of the neuron) and the sheath formed by oligodendroglia cells (neurilemma or Shvann's cells). They distinguish the *myelinated* nervous fibers (they also contain the myelinated membrane in addition to the axial cylinder, neurilemma, and the basal membrane), these fibers dominate in the somatic nervous

system; the *unmyelinated fibers* (they consist of the axial cylinder, neurilemma and basal membrane) form the autonomic (vegetative) nervous system mainly. The rate of conducting impulse in the myelinated fibers is much higher than in the unmyelinated, and it is about 120 m/s.

In the central and peripheral nervous systems the nervous fibers are closely located with each other carrying out different functions and providing conduction to many structures in different directions of the nervous system; that needs isolation of impulses from each other. This isolation is provided by the myelinated sheaths and **neuroglia** (a set of astrocytes, oligodendrocytes and microglical cells).

The function of the neuroglia ensuring the normal functioning of the nervous cells lies in isolation of the nervous fibers to carry out the mechanical, supporting, differentiating, trophic, protective and secretory functions, the regulating influence on the ionic structure and the nervous cells metabolism, the active participation in the higher brain functions and synthesis of mediators of the CNS.

Synapses are the specialized structures providing conduction of the nervous impulse from one neuron to the other. As a rule, they are formed between axons of one cell and dendrites of another (see the Figure 1.2 on the coloured insert).

In the structure of the synapse they distinguish *the presynaptic and postsynaptic parts and the synaptic cleft.* The presynaptic part is formed by the terminal branch of the axon transferring impulse of the nervous cell. It is covered with a presynaptic membrane and contains vesicles filled with mediators (biologically active agents — acetylcholine, norepinephrine). The postsynaptic membrane has a special protein — the mediator receptor. The synapse provides conducting of the nervous impulse only in one way (according to the law of the dynamic or physiological polarization of Ramyny Cajal's nervous cell). As for the functional features they distinguish two types of synapses: *stimulating*, i.e. promoting generation of impulses, and *inhibitory*, or capable to terminate the action of signals.

MAIN STAGES OF DEVELOPMENT OF THE NERVOUS SYSTEM

Functioning of the nervous system depends on the body reactivity, i.e. its ability to perceive irritation and to react on it by certain, in particular, the motor reaction. The compound morphological and functional features of the nervous system were created as a result of a long evolution, during which they were able to mark out schematically the following **stages of development**:

— The stage of the diffuse, reticular or asynaptic nervous system;
— The stage of the nodular or ganglionic nervous system;
— The stage of the tubular nervous system.

For the first time the nervous system appeared in *hydroid polyps* in the form of a network of epithelial cells and was called asynaptic, as it could conduct stimuli diffusively in all directions, without synapses (the reticular or diffuse nervous system) and provided the global reflex reactions. The nervous system of *worms* was created on the ganglionic type: symmetric, with two chains of ganglia (nodes) consisting of the nervous nodes cells and fibers. This system is synaptic; it is characterized by the ability to conduct stimuli only in one direction; it provides the differentiated adaptive reactions. The presence of

the pharyngeal node in worms indicates the origin of a primitive brain. In *mollusks* the nervous system is formed by the ganglionic type (as a network of the nervous fibers, which begin with the paired nodes). The tubular nervous system for the first time appears in *vertebrates;* it develops from the ectoderma; it is constructed by the segmentary type and equipped with the skeletal motor apparatus. So, fishes have already the spinal cord and the brain stem. The corpus striatum of *birds* reaches larger sizes; it is a substrate of the higher brain functions. In *mammals* the cortex was formed; in the *human beings* it reaches the highest development as the principal organ of thinking, speech and complex activity.

During ontogenesis the nervous system repeats all stages of phylogenesis. At first the cerebral (medullary) lamina is formed from the external ectodermal layer. Its edges are connected together forming the neural tube; the spinal cord is formed from its posterior part, and from its anterior part the brain is formed. Because of irregular growth of the anterior parts of the cerebral tube, the brain vesicles are formed; and respectively the anterior *(prosencephalon),* middle *(mesencephalon)* and posterior or rhomboidal *(rhombencephalon)* brain is formed as a result. This stage was called the stage of 'three vesicles'.

With time the terminal brain *(telencephalon)* was formed from the anterior part of the brain; it consist of the cerebral hemispheres, basal ganglia and intermediate brain *(diencephalon)*. Intermediate brain was formed by the following structures: thalamus, epithalamus, hypothalamus, metathalamus, optical path ways and nerves, and retina. This stage was called the stage of 'five'. The vesicles tectum and cerebral peduncles are formed from mesencephalon. From the posterior part the pons, cerebellum and medulla oblongata are formed.

From the posterior part of the spinal cord, forms and from neural the cavity of this tube the central channel of the spinal cord begins.

In the telencephalon there are the lateral ventricles; in the diencephalon there is the third ventricle of the brain; in the midbrain the aqueduct of mesencephalon is situated; it connects the third and fourth ventricles. The fourth ventricle is localized in *metencephalon.*

Formation of extremities in the course of evolution led to emergence of enlargements in the spinal cord: cervical — for the upper extremities (formed by the C_5—T_1 segments) and lumbar — for lower extremities (formed by L_1—S_2 segments).

Thus, during evolution the nervous system passes some stages, which are important for its morphological and functional development. They distinguish such **morphological stages** as:

— Centralization of the nervous system;
— Kefalization (from Greek *kephale* — the head);
— Corticalization (in *chordates*);
— Emergence of symmetric hemispheres (in the *highest vertebrata*).

GENERAL PRINCIPLES OF FUNCTIONING OF THE NERVOUS SYSTEM

During evolution there was a gradual centralization of the nervous system, which consisted in formation in the brain the centers subordinating the below-located structures to them. As a result the vital centers of the automatic regulation of different functions were created in the brain stem.